13 cont

4) a sequence derived from a sequence defined in 1), 2) or 3) by substitution, deletion or addition of one or more nucleotides with the proviso that said sequence still codes for said protease.

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4. (Amended) A nucleic acid sequence encoding the aminoacid sequence represented in Figure 2 (SEQ ID NO:6).

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6. (Twice Amended) An aminoacid sequence according to claim 5 characterized in that either it contains the sequence such as represented in Figure 2 (SEQ ID NO:6), or the amino acid sequence of Figure 2 (SEQ ID NO:6) modified by deletion, insertion and/or replacement of one or more amino acids with the proviso that such aminoacid sequence has the calpain activity involved in LGMD2 disease.

S/O G14

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15. (Amended) A method for detecting of a predisposition to a LGMD2 disease in a family or a human being, such method comprising the steps of:

- selecting one or more exons or their flanking sequences of the gene,
- selecting primers specific for these exons, or their flanking sequences, or an hybrid thereof,
- amplifying the nucleic acid sequences with these

primers, the substrate for this amplification being the DNA of a human being; and

- comparing the amplified sequence to the corresponding sequence derived from Figure 2 (SEQ ID NO:5, SEQ ID NO:68 and SEQ ID NO:69) or Figure 8 ((SEQ ID NO:1-SEQ ID NO:4).

16. (Amended) The method according to claim 15, characterized in that the primers are those selected from the group of:

a) those described in Table 1 (SEQ ID NO:10-SEQ ID NO:17);

b) those described in Table 3 (SEQ ID NO:18-SEQ ID NO:67); and

c) those including the introns-exons junctions of Table 2;

d) those derived from the primers in a), b), or c).

18. (Amended) A kit for the detection of a predisposition to LGMD2 by nucleic acid amplification characterized in that it comprises primers selected from the group of:

a) those described in Table 1 (SEQ ID NO:10-SEQ ID NO:17);